MYELOMALACIA, WITH ESPECIAL REFERENCE TO DIAGNOSIS AND TREATMENT.*

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Softening of the spinal cord, not consecutive to a myelitis, is practically always a sequel to arterial thrombosis.

It is an event of much more frequent occurrence than primary myelitis.

Early recognition of the nature of the process is of great practical importance, since upon such recognition depends the good effects of a rational therapy and consequently the best interests of the patient.

The foregoing propositions have been familiar to neurologists the world over for almost a decade, and have gradually gained in favor, until at the present day it is conceded that myelomalacia is a very frequent condition as compared with primary myelitis. In other words, a large majority of the cases on record as acute myelitis have been really cases of acute softening due to thrombosis and followed by inflammatory exudate around the softened area.

Williamson, in a remarkably comprehensive series of articles, (*Manchester Medical Chronicle*, 1894 and 1895) is usually credited with being the first to draw attention to the occurrence of softening in the cord of thrombotic origin, and to present actual demonstration of the thrombi in a case of myelitis as then recognized.

Bastian however (Quain's "Dictionary of Medicine," 1882 and 1885) had previously on purely rational grounds advocated the view that thrombotic spinal softening was a frequent event.

Singer (*Brain*, 1902. p. 332) has presented a very concise and satisfactory statement of the subject, and advocates the view that the "majority" of recorded cases of so-called "acute myelitis" are "not inflammatory" but due to thrombosis of spinal vessels.

While this frequency of myelomalacia and the corresponding rarity of true primary myelitis are generally recognized by neurologists, it is still an obvious fact that most of our standard textbooks on neurologic medicine, even those written by eminent masters of the art, have continued to group both conditions under the label "myelitis."

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In fact the term myelitis has about as much pathologic significance at the present day as the old term "apoplexy" in respect to brain lesions. It is not a pathologic diagnosis but merely a statement of loss of function in a given region of the nervous system.

Dana, for instance ("Text-book of Nervous Diseases," 5th Ed., 1901, pp. 249-250), states "It (myelitis) may be initiated or accompanied by a hemorrhage or softening from thrombosis of arteries, and the latter is often the case."

"*Nor can we clinically distinguish* between the cases due to a primary infection and those due to a hemorrhage and softening. Hence acute transverse myelitis may mean an acute inflammatory process or an acute softening." (Italics by present writer.)

Church and Peterson ("Nervous and Mental Diseases," 4th Ed., 1903, p. 351) consider "Thrombotic Softening of the Cord" under a separate heading. Here we find the statement that "Thrombosis of the arteries of the spinal cord undoubtedly occurs."

"This result has ordinarily been confounded with myelitis *and clinically presents the same picture.* (Italics by present writer.)

"As the *symptoms, treatment and prognosis are those of myelitis* it is not necessary to repeat them here."

Oppenheim ("Diseases of the Nervous System," Trans. by Mayer, 1900) includes under "Myelitis," "inflammations and softening processes" and further remarks "still, our conception of the term is not a precise one."

Even so discriminating an author as Gowers does not recognize myelomalacia as a clinical entity but speaks of "softening" only as a sequel of myelitis ("Manual of Nervous Diseases," Gowers and Taylor, 3d Ed., 1899).

Starr ("Organic Nervous Diseases," Phila., 1903) considers myelomalacia and myelitis separately, emphasizes the comparative frequency of myelomalacia and the rarity of myelitis, but states (pp. 347-348), "Whether a clinical distinction can be reached between these two types on account of the development in the infectious type of a marked febrile invasion with symptoms of an acute infection and leucocytosis, which are wholly absent in the second (myelomalacia) type, is a matter for further investigation."

The present paper is an attempt to establish on a practical basis, such a clinical differentation between the two diseases and to point out the principles of treatment appropriate to each.

DIAGNOSIS BETWEEN MYELOMALACIA AND PRIMARY MYELITIS.

Omitting consideration of questions of heredity and predisposition, as too remote for our present purpose, *the immediate prodromata* as a rule differ decidedly in the two conditions.

In myelomalacia the patient has commonly been attending to his customary work or duties, often with *no sign of illness before the onset* of the disease. There is no "chill." Pain is absent. There may be however transient attacks of numbness, tingling or sense of weight in one or more extremities, which are not of sufficient severity to prohibit his ordinary activities. Evidence of syphilis or other cause of vascular disease is commonly present.

In myelitis, on the contrary, the patient has been ill with some acute infection due to staphylococci, gonococci, pneumococci, etc., or one of the exanthemata is present. Traumatism, tumor or vertebral disease must of course be considered. In other words *the patient has been otherwise ill* aside from the cord disturbance proper.

The onset of myelomalacia is sudden as it is always in vascular nervous lesions whether the cord or brain be involved. There is no "chill"; pain is absent. Notwithstanding this suddenness of onset, the immediate loss of function may be slight in extent, perhaps a mere numbness or sense of weight in foot or leg, but it *persists* and *increases* often by additional *sudden increments* within a few hours or days. If these increments be absent, the symptoms, whatever their nature, remain stationary for a few days, perhaps a week and then show a period of extension or increase due to *exudative inflammation* around the softened area.

It is this secondary inflammation which often causes the patient to cease work on account of the motor weakness at this period.

Should the focus of softening, as is often the case, be confined to the irrigation field of one artery, say in an anterior horn, the resulting motor paralysis at the end of a week is still unilateral, while *marked sensory defects,* especially in recognition of heat and cold are present in the *opposite limb.*

In myelitis on the contrary the *onset is gradual.* Malaise, backache and "chill" often precede the sensory and motor symptoms; a convulsion may replace chill in the young. The primary cord symptoms are more commonly bilateral as well as more diffuse longitudinally. They steadily progress, (not by sudden increments) so that in a few days or a week, complete paraplegia is the rule.

Fever is commonly absent early in myelomalacia, and when it does develop later is due to some complication on the part of the intestinal tract or the bladder or to bed-sores.

In myelitis fever of moderate degree is commonly present at the start and if the cervical cord be involved a sharp rise of temperature to 105 or more is the rule.

As regards the transverse extent of the involvement of the cord, in myelomalacia this may be slight. Only one anterior horn area, of one segment, may be involved, one artery occluded. Sensory losses may be absent on the side of motor loss but marked in the opposite limb or area. This applies especially to temperature sensations. Such a "pattern" of functional losses means commonly, obstruction of a single anterior horn artery. But for its sudden onset it would simulate the "dissociation symptom" of syringomyelia.

In primary myelitis, an entire transverse section of the cord is involved within a few hours or days, and commonly several segments, as indicated by the diffuse character of the symptoms.

It is obvious that a general infection in the blood stream is not likely to be confined in its distribution to one artery.

Looking at the individual symptoms, we may consider: (1) Sensory defects, (2) motor paralysis, (3) changes in reflexes, (4) sphincter impairment, (5) bedsore.

In myelomalacia, which is usually of small extent at first, girdle sensation is often absent or may be unilateral. Anesthesia at the onset is less complete; often unilateral. Paresthesia, numbness and tingling are often present without absolute anesthesia. "Dissociation" of the thermal sense or of heat or cold separately is often present on one side of the body or in one limb, and this side is the one *opposite to the motor impairment.*

In myelitis "girdle sense" is commonly present, numbness and tingling are followed rapidly by absolute bilateral and complete anesthesia for all forms of sensation below the level of the lesion; "dissociation" is not present as a rule.

In myelomalacia, *motor symptoms* are commonly *unilateral* at the beginning, affecting a limb or a muscle group. Muscular atrophy and R.D. correspond, and are often less extensive than the initial symptoms would lead us to expect, owing to re-establishment of circulation in part of the affected territory by contiguous vessels. In case of involvement of another vessel on the opposite side, the "level" often does not correspond as shown by the "pattern" of motor, sensory and trophic defects.

In myelitis, *paralysis* is commonly *bilateral, symmetrical,* and *complete* below the lesion in a few hours or days. Muscular atrophy and R.D. are bilateral and symmetrical in the territory supplied by the affected segments.

The *reflexes* in myelomalacia are commonly unlike on the two sides of the body. One knee-jerk may be gone, in lumbar lesions, the other simply diminished, sometimes accentuated.

The *plantar reflexes* are likewise apt to be *unlike,* e.g., a Babinski toe extension on the paralyzed side and a flexion or simply rigidity in the opposite foot.

In myelitis the knee-jerks are at first abolished or markedly diminished on *both sides,* later becoming accentuated if the mid-lumber segments be intact and the main lesion be not a "complete" one, which it practically seldom is. The *planter reflexes* at first as a rule are *also abolished;* a double "Babinski' toe extension appearing later.

Sphincter Control.—In myelomalacia, the *sphincters may escape* or may be affected for a few hours or days only.

In myelitis *complete prolonged loss* of sphincter control is the rule.

Bed-sores are often absent in myelomalacia and may be prevented entirely in most cases by care and cleanliness. The rarity of bedsores may possibly be due to the preponderance of the lesion in the anterior horns, circumscribed affections of which, as is well known, seldom or never cause bedsores.

In *myelitis,* on the contrary, *bedsores appear early* and are seldom preventable by any degree of care.

The absence of leucocytosis would be expected in myelomalacia; its presence in myelitis, as indicated by Starr.

The foregoing remarks on diagnosis may be tabulated as follows:

	MYELOMALACIA.		MYELITIS.
1.	No preceding disability generally.	1.	Preceding disability, injury or acute illness present.
2.	Onset of paralysis sudden.	2.	Onset of paralysis gradual or rapid, not sudden.
3.	Fever absent, pulse low-tension.	3.	Fever present, pulse high tension early in the disease.
4.	Chill absent.	4.	Chill sometimes present.
5.	Rigidity of spine absent, Spasm of extremities absent. Area affected variable often unilateral and slight in extent. Paralysis often monoplegic. Extension by sudden increments.	5.	Rigidity of spine may be present. Spasm of extremities often present. Area affected usually one or more entire segments. Paraylsis always paraplegic. Extension steadily progressive.
6.	Dissociation symptom (Thermo-anesthesia) often present on non-paralyzed limb or on both limbs in paraplegic cases.	6.	Dissociation symptom absent. Anesthesia bilateral and symmetrical to all sensations below lesion.
7.	Girdle symptom often absent.	7.	Girdle symptom present.
8.	Knee-jerks unlike, often unilaterally abolished.	8.	Knee-jerks abolished or diminished bilaterally and equally.
9.	Plantar reflexes unlike; unilateral "Babinski" sign common.	9.	Plantar reflexes abolished early on both sides (double "Babinski" sign later).
10.	Sphincteric control may remain of may be lost for a few days only.	10.	Sphincteric control abolished for a prolonged period.
11.	Bedsores often absent; usually so in small discrete lesions.	11.	Bedsores inevitable.
12.	Leucocytosis absent.	12.	Leucocytosis probably present.

THERAPEUTIC CONSIDERATIONS:

Early recognition and prompt treatment render the prognosis of myelomalacia much more favorable than that of myelitis. The main indications for treatment are obviously the same as they are in acute cerebral softening from thrombosis, namely:

1. To increase the blood pressure and encourage the peripheral circulation.

2. To promote liquefaction and limit organization in the inflammatory exudate which surrounds the softened area after a few days.

By meeting the first of these indications we improve the circulation not only in the territory adjacent to the blocked vessel, but likewise in vessels elsewhere, which may at that moment be undergoing a gradual occlusion. We hold the way open for the blood stream, by cardiac stimulants and arterial dilators, while our slower acting "alteratives" or "exudate destroyers," as mercury and the iodides should be called, are attacking the enemy already entrenched. By these measures much of the consecutive cicatricial pressure in the vicinity of the actual lesion is prevented and ascending and descending degenerations reduced to a minimum.

Rest on an air mattress with frequent changes of posture, extreme cleanliness, proper attention to the bladder and rectum, and after the acute stage, passive motion and massage of the paralyzed limbs; galvanism to the atrophied muscles and general hygienic measures complete the treatment. The results of such a plan of treatment must obviously average better than those of a merely "expectant plan."

The treatment of myelitis does not call so urgently for cardiac stimulants or arterial dilators; in fact the conditions may even furnish indications for opposite measures.

The spinal rigidity and spasms in the limbs, both of which are more apt to occur in myelitis than in myelomalacia, are best controlled by sulphonal in my experience, though bromides and chloral are also effective.

The "antiphlogistic" or "exudate destroying" effect of mercury and the iodides is not to be ignored in myelitis, even when syphilis is not present.

ILLUSTRATIVE CASES:

CASE I. Myelomalacia:

An active business man of 35, well-developed and of good habits, consulted me in reference to temporary loss of power in his legs a few days previous. There had been some premonitory numbness for a few hours and his feet felt "heavy." On his way home he had reached his premises when he suddenly sank to the ground, seemingly from weakness in both legs. There was no giddiness or mental confusion. He had not been ill nor drinking; nor had he experienced any injury or shock. He could not rise for some minutes, but did not call for assistance and finally managed to reach the house and retire to bed. He found it difficult to turn over while in bed, and on attempting to get up, he found he could only do so by rolling out and assisting himself with his hands. With some effort he managed to get about his business, feeling a sense of weakness in the legs.

After four days he consulted an oculist about some defect in vision and an exudative choroiditis was recognized. By reason of the history of weakness in legs he was referred to me for opinion and advice.

I found a healthy looking man, well-developed and nourished, who denied any knowledge of syphilis, used no alcohol, but smoked freely.

His gait was not visibly affected, but he still complained of an indefinite weakness in the legs. There was no "girdle" sensation. Pulse 108, very low tension, small volume. Sensation appeared normal to tact and pain everywhere, but an area of the outerborder of the left foot, extending from ankle to root of toes and nearly to the median line of the dorsal surface was insensitive to heat and cold.

The knee-jerks were unequal, the right about normal, the left decidedly diminished.

The plantar reflexes were also unlike, the right showing flexion and extension alternately, with occasional rigidity in a midway position, the left sharply flexor at all times.

A test for Oppenheim's reflex (by steady deep stroking at inner border of tibia) resulted in developing a distinct constant extension of the right great toe with flexion of the others. On the left foot the response was flexion of all toes.

The sphincters were not affected. Bedsore absent. Blood examination not made.

A diagnosis was made of thrombosis of a minute vessel, supplying portions of the IV and V lumbar segments of the right side.

The patient was put to bed, strychnia, nitroglycerin and strophanthus given to improve the circulation and mercury administered by hypodermic injection of "gray oil," alternating with iodopin given in the same manner.

The pulse improved, the sense of weight in legs and feet disappeared in a few days. In about two weeks the knee-jerks were alike and normal and the plantar reflexes were also normal and equal.

The patient has been well for eight months, with no further vascular accidents, and general good health.

CASE II. Myelomalacia:

A man of 40, colored, shop porter. Complete loss of power in right arm of sudden onset while sleeping. Three days later was admitted to my service at the Cincinnati Hospital.

Condition on Admission.—Man of average height, good general development. Habits good as to alcohol. Syphilis probable some years previous. Mental condition good. No speech defect.

Motion.—(See Fig. 1.)—Examination showed absolute flaccid paralysis of entire right upper extremity; even the scapula could not be moved by any voluntary effort. Is not conscious of any defect in left arm and uses it freely.

Tested by Matthieu dynamometer, however, the grasp in the left hand is only 55 *K* (estimated to be about 60 per cent. of normal). No facial weakness evident at this examination (see later). No complaint of leg weakness. Gait is good.

Sensation.—No subjective sense of pain, no dizziness, no mental confusion at onset or since. No loss of tactile or pain sense anywhere. Sense of heat and cold lost, or confused, in irregular area over the right shoulder anteriorly, over right pectoralis major and over outer half of right arm and forearm anteriorly to a point three inches above wrist. Cannot recognize cold with right hand or fingers. Cannot recognize heat or cold with left hand and over a longitudinal strip on posterior external surface of left arm and forearm. (See Fig. 2.)

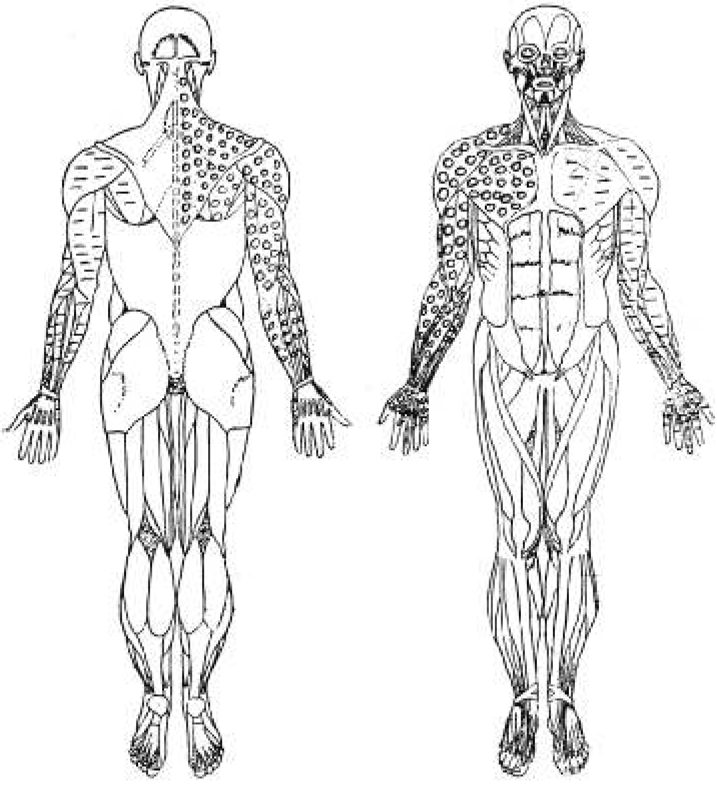

Fig. 1. Chart showing motor defects. Circles indicate absolute loss of power; dashes, partial loss.

Reflexes.—Pupils do not dilate in darkness or on irritation of skin of neck; they respond well to accommodation and convergence.

The supra-orbital reflexes are present and active right and left. Jaw-jerk absent. Elbow jerk present right and left. Wrist and supinator jerks present right and left. Hypochondriac reflex, right absent, left present clear and sharp. Inguinal reflexes present, right and left equal. Cremasteric reflexes absent. Knee-jerks absent right and left. Front tap contraction absent right and left. Tendo-Achilles jerks absent right and left. Plantar reflexes: Right, extension of great toe and flexion of all the smaller toes; a good "Babinski" response. Left, rigidity of great toe, with occasional slight extension; flexion smaller toes, incomplete "Babinski" response.

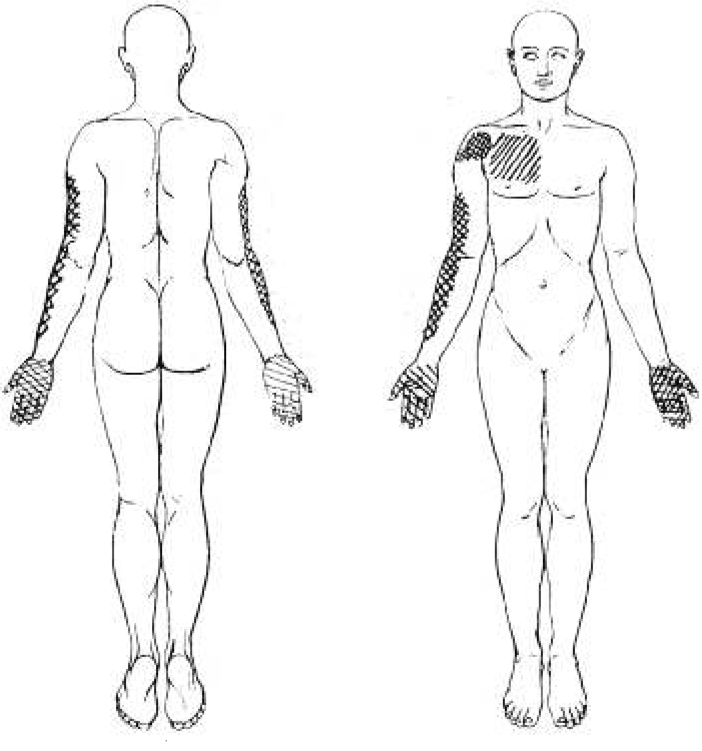

Fig. 2. Chart showing sensory defects.

Lines sloping from left to right—Loss of cold sense.
Lines sloping from right to left—Loss of heat sense.
Lines crossed—Loss of both heat and cold.

No sphincter defect. No trophic lesion. No obvious wasting. Pulse 96, low tension; temperature 98, respiration 16.

Seven days after the onset of the arm palsy (four days after admission to hospital) paralysis of right lower face and of the right orbicularis palpebrarum developed. The mouth was drawn to left, the tongue protruded to the right (actually, not apparently).

Electrical examination of the right arm and hand muscles at this date showed sluggish contraction to galvanism with polar reversal (A.C. > K.C.) in deltoid, and in thena and hypo-thenar groups, quick contraction with K.C. > A.C. in right trapezius, biceps, wrist flexors and extensors. Some wasting in arm was evident. The patient made a gradual improvement so that in two weeks he could flex the elbow slightly. One month later slight improvement manifest in flexion at elbow.

Five weeks later all movements of right arm and shoulder could be performed weakly, and patient could raise hand to top of head. Grasp of right hand registered 25 as a maximum, while the left registered 45%. (Reduction in power estimated at 75% in right grasp and 50% in left grasp.)

Unfortunately the patient left the house during my absence after a change of internes and the conditions of knee-jerks or pupils and other important items were not noted.

To sum up the conditions present in this case;—sudden onset, flaccid paralysis, absence of associated shoulder movements, the distribution of cutaneous sensory defects, the bi-lateral but unsymmetric character of the motor and sensory symptoms, the absence of mental confusion, or speech defect all point to a spinal location of the lesions. The partial recovery of function would indicate multiple minute areas of softening due to gummatous infiltration and thrombosis of numerous small vessels in the cervical cord from the III to VIII segments, rather than a single extensive lesion.

CASE III. Myelitis (primary):

As an example of true primary myelitis I cannot do better than refer to an abstract of the case reported at this meeting by my friends, Drs. Zenner and Wolfstein, through whose courtesy I had the privilege of studying it during life and afterward.

Man of 33, driver; general good health, gonorrhea within six months, followed by "la grippe." Syphilis not probable. Two weeks previous to coming under Dr. Zenner's observation his foot slipped in mounting his wagon step and he stepped forcibly to the ground. He went on with his work, but in two hours noted a numb feeling in his left side. The next day numbness in left leg and slight difficulty in walking. The right leg was affected similarly in a week. Then the right arm weakened and he was compelled to quit work. Three days later he lost control of the sphincters.

Three days after this Dr. Zenner saw him. He was completely paraplegic.

Temperature 101, pulse 108. Temperature during next twenty days varied between 100 and 102 usually, once rising to 104.

The paralysis extended and involved both arms and was preceded by spasmodic movements in right arm and hand. Dyspnea and dysphagia appeared. The knee-jerks disappeared; the right going first. The plantar reflexes were absent when patient was first seen. A "bedsore" developed over heel, four days after coming under observation (eighteen days after onset). Thirty-one days after onset, stupor, twitching of face muscles. Pulse 63, respiration 30. Death on the thirty-fourth day.

A diffused round-celled infiltration with no gross vascular lesion was found extending from the lumbar to the mid-cervical region. (See this JOURNAL, p. 190.)

As regards the relative frequency of myelomalacia and myelitis based on clinical observation, the following figures may be of some interest:

They are the results of a review of the neurological service in a large municipal hospital (The Cincinnati Hospital) for the past five years, lacking four months.

During this period 27,397 patients were treated in the hospital, of which 1,649 were admitted to the neurological service.

I have also tabulated separately the cases seen by me in private practice, and of which I have notes, during the same period.

Reviewing both sets of cases of "paraplegia" "myelitis," etc., during this period, I would tabulate them in accordance with present generally accepted views as follows:

				FINAL RESULT			
Hospital cases—	Number.	Males.	Females.	Practical Recovery.	Improved.	Died.	Not Stated.
•Myelomalacia (syphilitic)......	25	19	6	..	17	4	4
Myelomalacia (senile).........	2	1	1	..	1	1	
Myelomalacia (typhoid)........	2	1	1	..	..	1	1
Private cases—							
Myelomalacia (syphilitic)......	14	13	1	4	8	2	
Myelomalacia (alcoholic?).....	2	2	..	..	1	1	
Myelomalacia (la grippe)......	1	1	..	..	1		
Myelomalacia (puerperal).....	1	..	1	1			
Myelomalacia (senile).........	1	..	1	..	1		
	—	—	—	—	—	—	—
Hospital cases—	48	37	11	5	29	9	5
Myelitis (traumatic)...........	5	3	2	1	..	2	2
Myelitis (compression caries).	2	2	..	1	..	..	1
Myelitis (infectious)...........	4	3	1	..	..	1	3
Private cases—							
Myelitis (traumatic)...........	4	3	1	..	3	1	
Myelitis (gunshot)............	3	2	1	..	1	2	
Myelitis (compression caries).	1	1	..	..	..	1	
Myelitis (sarcoma)............	1	1	..	..	..	1	
Myelitis (hematorrhachis trau.)	1	1	..	1	..		
	—	—	—	—	—	—	—
	21	16	5	3	4	8	6